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Case Report

A rare case of elephantiasic pretibial myxedema with secondary cutis verticis gyrata

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ABSTRACT

A 61-year-old man with a history of thyrotoxicosis, diabetes mellitus, hypertension, and dyslipidemia presented to the dermatology outpatient clinic with bilateral non-pitting edema, asymmetrical yellowish-brown plaques, and nodules associated with some areas of hyperpigmentation, Peau'd orange appearance and elephantiasis-like skin lesions over both low legs. It was diagnosed as pretibial myxedema attributed to thyrotoxicosis. Additionally, convoluted folds and deep furrows similar to the cerebral cortical surface were seen on the forehead suggesting secondary cutis verticis gyrata (CVG) and our approach to the patient will be discussed in this case report.

Keywords: Pretibial myxedema, Thyroid dermopathy, Thyrotoxicosis, CVG

INTRODUCTION

Pretibial myxedema, also known as thyroid dermopathy, is found in 0.5-4.3% of thyrotoxic patients.[1] There are four types of pretibial myxedema: non-pitting edema with typical skin color changes (43.3%), plaque type (27%), nodular type (18.5%), and elephantiasic type (2.8%).[2] It is noted that elephantiasic type is the least common form of pretibial myxedema. CVG is a rare disorder characterized by convoluted folds and furrows resembling the surface of the cerebral cortex. It has an estimated prevalence of 1 in 100,000 of the male population and 0.026 in 100,000 of the female population.[3] It can be classified into primary CVG and secondary CVG.[3] It usually occurs over the scalp but rarely, other body parts, e.g., forehead and face, can be affected.[4] There are only a few case reports regarding the association between CVG and endocrine diseases. In our case, we will present two rare clinical presentations of a thyrotoxic patient: elephantiasic type pretibial myxedema and secondary CVG on the forehead.

CASE REPORT

A 61-year-old Thai man with underlying diseases including diabetes mellitus, hypertension, and dyslipidemia came to the outpatient department of Bangphai hospital, Bangkok with the chief complaint of palpitation and fatigue five years ago. He was diagnosed with thyrotoxicosis; treated and maintained with radioactive iodine and methimazole respectively. Three years ago, he presented with dry irritated eyes and blurred vision, which was diagnosed as thyroid ophthalmopathy, and underwent orbital decompression surgery. Six months later, swellings developed in the legs from toes to shins with dark-brown discoloration over the lesions. The latter became gradually extensive causing functional limitations with walking. However, he denied pruritus, pain, ulceration, or swelling in other parts of the body.

After one year, he came to the general medicine outpatient department for his follow-up and was referred to our dermatology department. On physical examination,

we noticed bilateral, frim, non-pitting edema with asymmetrical dark brown plaques and nodules (2×3 cm) predominantly on shins and dorsum of the feet extending to calves with thickened, harden, and elephantiasic skin areas. Peau d'orange appearance was also noted with some yellowish to dark brown discoloration (Figure 1). Then, the diagnosis of elephantiasic pretibial myxedema was made, and a skin biopsy was taken. The pathology report showed fibrous tissue components and some inflammatory cell infiltration; others were unremarkable. He was treated with intralesional corticosteroid injection (Triamcinolone (10 mg) 1 ml: Normal saline 1 ml) on each side of the leg every two weeks.

In addition, there were vertical deep linear furrows with hypertrophy and skin folds on his forehead. (Figure 2 and 3). It was diagnosed as secondary CVG and managed conservatively.

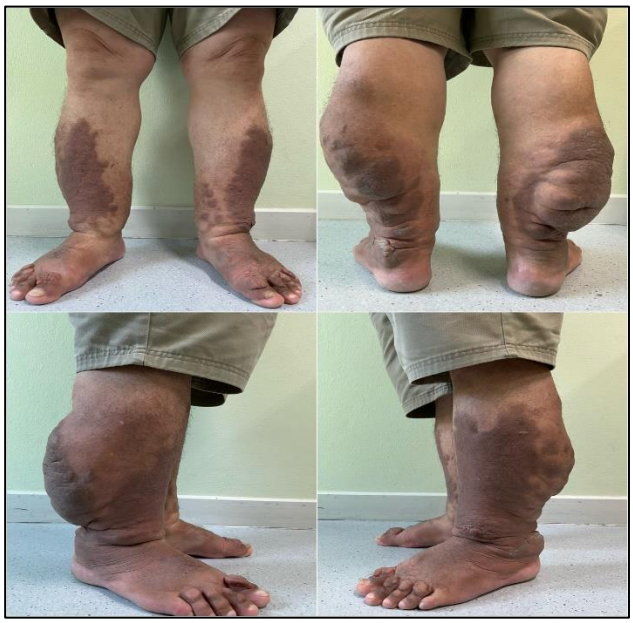

Figure 1: Lesions on the legs of the patient: elephantiasis pretibial myxedema.

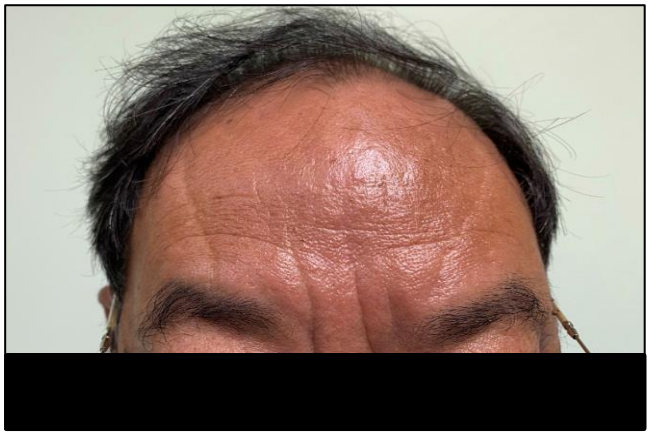

Figure 2: Lesions on the forehead of the patient: CVG (Front view).

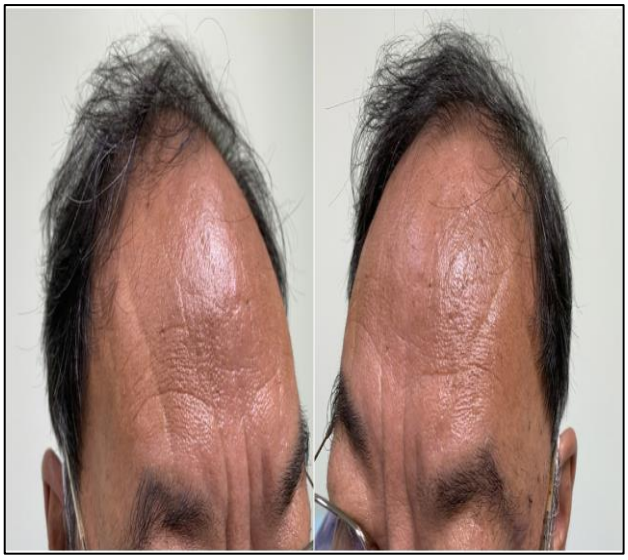

Figure 3: Lesions on the forehead of the patient: CVG (Side views).

DISCUSSION

Pretibial myxedema is a rare presentation of thyrotoxicosis with an estimated prevalence of 5 % in patients with Graves' disease. But it is more common in patients with ophthalmopathy (15%).[5] In this case, we can see the association of ophthalmopathy with dermopathy. Pathogenesis is supposed to be the accumulation of hydrophilic glycosaminoglycans (GAG) which causes fluid collection in dermal tissues leading to compression and subsequent occlusion of small lymphatic vessels with resultant edema. Dermal fibroblasts normally express TSH-receptors proteins ligated by TSH-receptor antibodies and/or antigen-specific T cells. Cytokines released in lymphocytic infiltration stimulate fibroblasts to produce GAG.[1]

Pretibial myxedema usually presents as bilateral, asymmetric, non-pitting, raised waxy lesions, usually flesh color or yellowish-brown appearance. Areas of hyperpigmentation, hyperkeratosis, and local hyperhidrosis may be present. It can be indurated with prominent hair follicles resembling an orange peel appearance which is called Peau d' Orange.[1] Some cases can completely progress to legs, feet, or hands and become an elephantiasis form characterized by thickened, woody, firm areas with non-pitting edema, skin fibrosis, and verrucous nodules. The patient presented with typical lesions including pretibial myxedema with hyperpigmentation, Paeu d' orange and elephantiasic lesions (Figure 1).

Pretibial myxedema is usually a clinical diagnosis. The biopsy is normally only done in patients with skin lesions lacking active hyperthyroidism or a history of autoimmune thyroid disease. Histopathology usually shows mucinous edema and fragmentation of collagen fibers with deposition of acid mucopolysaccharides in the

papillary and reticular dermis, with subsequent extension into deeper tissue. Increased numbers of fibroblasts can also be seen.[6] In our case, a punch biopsy was taken for diagnosis, and it showed fibrous tissue components and some inflammatory cell infiltration.

Majority of patients with pretibial myxedema do not need treatment. Treatment is only indicated for cosmetically objectionable lesions and is usually based on case reports, case series, and clinical experiences.[1] Topical corticosteroids under occlusive dressing (or) intralesional corticosteroids are usually prescribed as a primary treatment.[7] There are some case reports with IV IgG, plasmapheresis, and injections of octreotide in conjunction with surgery.[1] In our case, we gave an intralesional corticosteroid injection every 2 week. However, it is said that longstanding pretibial myxedema may be quite resistant to all therapy and most server disease (elephantiasic type) are less likely to have remission despite treatment with corticosteroids. Thus, we still need to monitor the long-term effect and adjust accordingly although we saw some improvements in follow-up.

Another clinical presentation in our case is secondary CVG. It is a rare condition characterized by skin lesions with convoluted folds and deep furrows resembling the cerebral cortical surface. It typically affects the scalp, but other body parts such as forehead and face may also be affected.[4] Our case presents a rare involvement of CVG on the forehead. CVG can be classified into primary CVG and secondary CVG;[3] the former includes essential and non-essential types. The essential form of primary CVG usually occurs in isolation in otherwise normal individuals whereas the non-essential form is often associated with neuropsychiatric or ophthalmologic abnormalities.[8] Secondary CVG is more common than primary CVG and usually associated with localized or systemic inflammatory or neoplastic disease, several genetic syndromes: Turner syndrome, Noonan syndrome, craniosynostosis syndromes, and endocrine diseases (e.g., acromegaly, myxedema, Graves' disease.[9] In our case, we present two rare clinical manifestations of thyrotoxicosis: pretibial myxedema and secondary CVG (Figure 2 and 3).

The pathophysiology of CVG is poorly understood and depends on underlying conditions. The role of proinflammatory cytokines, bacterial and mycotic superinfections, and genetics: autosomal dominant mutation in the fibroblast growth factor receptor 2 (FGFR2) gene have been described in the literature. Increased IGF levels, prominent presence of fibroblasts, and interstitial deposition of glycosaminoglycans are also known as supposed mechanisms for CVG.[9] Thus, this is supposed to be the mechanism of CVG in our case.

The diagnostic approach to CVG varies depending on history and physical examination. In primary CVG, a biopsy is usually not needed. In newborns with Turner syndrome/ Noonan syndrome, a skin biopsy is necessary if the lesion is suspicious of a cerebriform intradermal nevus because it can transform into malignant melanoma.[10] Biopsy is required to get a precise diagnosis of secondary CVG. Our case did not get a biopsy due to salient clinical features and the patient's refusal to get it from his face.

CVG is almost always benign and causes aesthetic problems rather than functional issues. There is no treatment with proven efficacy for CVG.[9] Therefore, focus-on therapy for primary CVG is good scalp hygiene to avoid the accumulation of secretion in furrows.[10] If extensive lesions are present, skin scalp subcision under local anesthesia may be considered. For secondary CVG, it is suggested to treat the underlying cause. Therefore, no specific treatment was given in our case and only focused on managing thyrotoxicosis and pre-tibial myxedema.

CONCLUSION

In conclusion, this is how we approach a rare case of elephantiasic pretibial myxedema and secondary CVG in a patient with thyrotoxicosis. The standard management of these conditions is still needed to be evaluated in more case studies and clinical trials in the future. Nevertheless, since the elephantiasic type of pretibial myxedema, as well as CVG, are rare clinical presentations in our clinical practice, the authors hope this case report will be beneficial for readers regarding the diagnosis and making therapeutic decisions of these two conditions in daily clinical practices of dermatology.

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